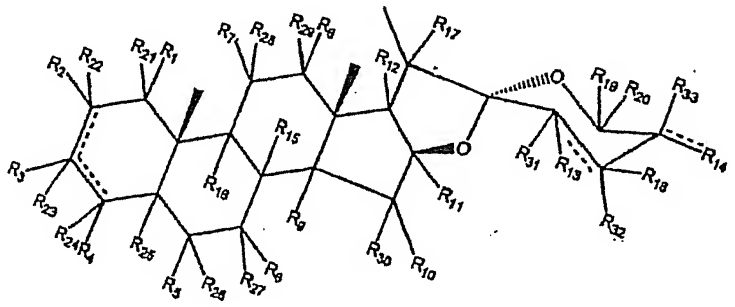
Amendments to the Claims:

This listing of claims will replace all prior versions, and listings, of claims in the application:

Listing of Claims:

1. (withdrawn) Use of one or more active agent selected from :

A. compounds of Formula I :



(I)

wherein in the general formula (I):

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me- SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R = alkyl or acyl group;

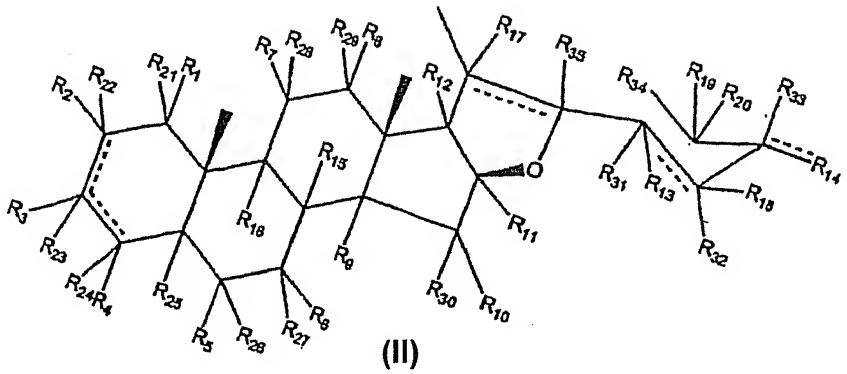
- R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33} can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R = alkyl or acyl group;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group;

B. compounds of Formula II :



(II)

wherein in the general formula (II) :

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10} R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}, R_{34} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

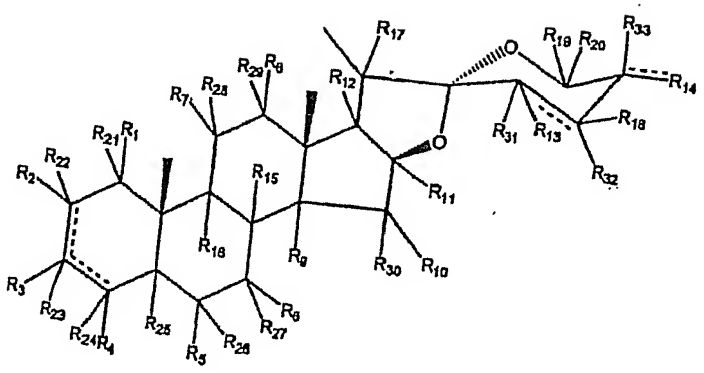
- R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33}, R_{35} can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group;

C. compounds of Formula III :



(III)

wherein in the general formula (III) :

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{14}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}, R_{33}, R_{34}, R_{35}, R_{36}, R_{37} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (ME-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

-R_9, R_{11}, R_{12}, R_{15}, R_{16}, R_{17}, R_{25} can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group, and

the stereochemistry of R_{25} is in the β orientation;

D. sapogenin derivatives bearing at least one X radical substituent,

wherein X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-),

-N_3-, NH_2-, $MeSO_2NH$-, and

-alkyl ; and

E. derivative forms of any of the above compounds, in which the carbon atom at the 3- position or, in the case of Formulae II and III, the 3-position carbon atom, the 26-position or each of the carbon atoms at the 3-and 26-positions, carries an O-sugar moiety wherein the sugar group is a mono-, di-or tri-saccharide;

all their stereoisomers and racemic mixtures, all their pharmaceutically acceptable pro-drugs and salts, and all mixtures and combinations thereof

in the treatment or prevention of, or in the preparation of compositions for the treatment or prevention of, (i) non-cognitive neurodegeneration, (ii) non-cognitive neuromuscular degeneration, (iii) motor-sensory neurodegeneration, or (iv) receptor dysfunction or loss in the absence of cognitive, neural and neuromuscular impairment, in human and non-human animals suffering therefrom or susceptible thereto.

2. (withdrawn) A use according to claim 1, wherein the active agent, or at least one of the active agents, is selected from:

a. Compounds of the above general formula I, wherein:

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}, are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me- SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R = alkyl or acyl group;

- R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33} can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R= alkyl or acyl group;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group,

and the stereochemistry of R_{25} is in the ß orientation;

b. Compounds of the above general formula I, wherein:

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me- SO-), (Me-SO_2), N_3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R = alkyl or acyl group;
- R_9, R_{12}, R_{15}, R_{16}, R_{17} = H,
- R_{11}, R_{14}, R_{25}, R_{33} can be either a H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl or absent or OR where R = alkyl or acyl group;

..... represents an optional double bond

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group,

and the stereochemistry of R_{25} is in the ß orientation;

c. Compounds of the above general formula I, wherein:

-R_1= R_2= R_4= R_5= R_6= R_7= R_8= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_Z) R_{22}= R_{23}= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32}= R_{33} = H,
- either R_{33} or R_{14} = CH_3

..... represents a single bond,

- the methyl group at C_{25} may be either in the R or S configuration

- the stereochemistry of R_{25} is in the β orientation and

wherein in addition to the above

at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O, and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and

- N_3-, NH_2-, $MeSO_2NH$- - alkyl ;

d. Compounds of the above general formula I, wherein:

-R_1= R_2= R_4= R_5= R_6= R_7= R_8= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21}= R_{22}= R_{23}= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32} = H,

-R_{14}= R_{33} = CH_3,

..... represents a single bond,

- the stereochemistry of R_{25} is in the β orientation and

wherein in addition to the above

at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O,

and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and - N_3-, NH_2-, $MeSO_2NH$-

-alkyl ;

e. Compounds of the above general formula II, wherein

-R_1, R_2, R3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31} R_{32}, R_{34} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO_2), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

R_9, R_{11}, R_{12}, R_{14}, R_{15}, R_{16}, R_{17}, R_{25}, R_{33}, R_{35} can be either a H, OH.. halo atom, (ME-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group,

and the stereochemistry of R_{25} is in the ß orientation;

f. Compounds of the above general formula II or carbohydrate derivatives thereof, wherein:

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me- SO-), (Me-SO_2), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

- R_9, R_{12}, R_{15}, R_{16}, R_{17} = H, - R_{34}= either H, OH, =O, and OR where R = alkyl, acyl or carbohydrate and

- R, R) $_4$, R_{25}, R_{33}. R_{35} can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group,

and the stereochemistry of R_{25} is in the ß orientation;

g. Compounds of the above general formula II or carbohydrate derivatives thereof, wherein:

-R_1= R_2= R_4= R_5= R_6= R_7= R_8= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21}= R_{22}= R_{23}= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32}= R_{33} = H,

-R_{14} = CH_3,

- R_{34}= -OH or -OR where R = alkyl, acyl or carbohydrate and

R_{35} = H or is absent

..... represents an optional double bond, and

- the methyl group at C_{25} may be either in the R or S configuration and

and the stereochemistry of R_{25} is in the ß orientation

wherein in addition to the above

at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O,

and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of:

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and

- N_3-, NH_2-, $MeSO_2NH$-

- alkyl ;

h. Compounds of the above general formula II or carbohydrate derivatives thereof, wherein:

- R_1= R_2= R_4= R_5= R_6= R_7= R_8= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21}= R_{22}= R_23= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32}= H,

-R_{14} = R_{33} = CH_3,

- R_{34}=-OH or-OR where R = alkyl, acyl or carbohydrate and

R_{35} = H or is absent

..... represents an optional double bond, and

the stereochemistry of R_{25} is in the ß orientation and

wherein in addition to the above

at least one of R_3 OR R_{23} is a X radical, the possible remaining substituent being H, OH, =O,

and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of:

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and

- N_3-, NH_2-, $MeSO_2NH$-

- alkyl;

i. Compounds of the above general formula III, wherein:

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{14}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}, R_{33}, R_{34}, R_{3s}, R_{36}, R_{37} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

-R_9, R_{11}, R_{12}, R_{15}, R_{16}, R_{17}, R_{25} can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or R_{14} = alkyl group, and

the stereochemistry of R_{25} is in the P orientation;

j. Compounds of the above general formula III or carbohydrate derivatives thereof, wherein:

- R_1, R_2, R_3, R_4, R_5, R_6, R_7, R_8, R_{10}, R_{13}, R_{14}, R_{18}, R_{19}, R_{20}, R_{21}, R_{22}, R_{23}, R_{24}, R_{26}, R_{27}, R_{28}, R_{29}, R_{30}, R_{31}, R_{32}, R_{33}, R_{35}, R_{36}, R_{37} are, independently of each other, either H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl or acyl group, or absent;

- R_9, R_{12}, R_{15}, R_{16}, R_{17} = H,

- R_{34} = H, OH, =O, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_2NH$-, alkyl, OR where R = alkyl, acyl or carbohydrate, or absent;

- R_{11}, R_{25}, can be either H, OH, halo atom, (Me-S-), (Me-SO-), (Me-SO_2-), N_3-, NH_2-, $MeSO_ZNH$-, alkyl, OR where R = alkyl or acyl group, or absent;

..... represents an optional double bond,

wherein in addition to the above

- either R_{33} or $R_{,4}$ = alkyl group,

and the stereochemistry of R_{25} is in the β orientation;

k. Compounds of the above general formula III, wherein:

-R_1= R_2= R_4= R_5= R_6= R_7= R_8= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{21}= R_{22}= R_{23}= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32}= R_{33} = H,

- R_{14} = CH_3,

- R_{34}= -OH or-OR where R = alkyl, acyl or carbohydrate and

R_{35} = H or is absent

R_{37}=H, or is absent

R_{37}=H, -OH or =0

R_{36}= H or-OH

..... represents a single bond, and

- the methyl group at C_{25} may be either in the R or S configuration and

the stereochemistry of R_{25} is in the β orientation

wherein in addition to the above

at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O,

and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and

- N_3-, NH_2-, $MeSO_2NH$-

- alkyl;

j. Compounds of the above general formula IN or carbohydrate derivatives thereof, wherein:

-R_1= R_2= R_4= R_5= R_6= R_7= R_{89}= R_{10}=R_{11}= R_9= R_{12}= R_{13}= R_{15} = R_{16} = R_{17} = R_{18} = R_{19} = R_{20} = R_{2L}= R_{22}= R_{23}= R_{24}= R_{25}= R_{26}= R_{27}= R_{28}=R_{29}= R_{30}= R_{31}= R_{32} = R_{19} = R_{20} = H,

-R_{L4}= R_{33} = CH_3,

- R_{34}=-OH or-OR where R = alkyl, acyl or carbohydrate and

R_{35} = H or is absent

R_{37}= H,-OH OR =O

R_{36} = H or-OH

..... represents a single bond, and

- the methyl group at C_{25} may be either in the R or S configuration and

the stereochemistry of R_{25} is in the β orientation

wherein in addition to the above

at least one of R_3 or R_{23} is a X radical, the possible remaining substituent being H, OH, =O,

and OR where R = alkyl or acyl group or absent,

and X is chosen from the group consisting of :

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-), and

-N_3-, NH_2-, $MeSO_2NH$-

- alkyl;

m. Substituted sapogenins wherein at least one OH-group of the sapogenin is substituted with X, chosen from the group consisting of:

- halo atom,

- (Me-S-), (Me-SO-), (Me-SO_2-),

- N_3-, NH_2-, $MeSO_2NH$-, and

- alkyl;

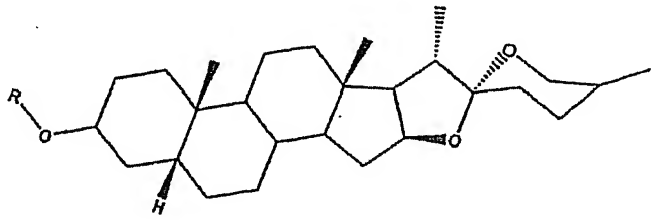
n. Sapogenins defined above wherein in the definition of X the halo atom is a fluoro atom;

o. Substituted sapogenins selected from:
(3 ß-fluoro-5ß, 20A, 22A, 25R-spirostane), (3, 3-difluoro-5 (3, 20A, 22A, 25R-spirostane), (3A-methylsulphonylamino-5 (3, 20a, 22A, 25R-spirostane), (3a-azido-5 (3, 20a, 22a, 25R-spiro- stane), (3a-amino-5 (3, 20a, 22a, 25R-spirostane), and their

stereoisomers and racemic mixtures, their pharmaceutically acceptable pro-drugs and salts;

p. Substituted sapogenins wherein the parent sapogenin which is then substituted with at least one X radical as defined above is selected from sarsasapogenin, episarsasapogenin, smilagenin, epismilagenin, and anzurogenin-D;

q. Compounds of the general formula Ia :



(Ia)

wherein the group R is selected from hydrogen; alkylcarbonyl; alkoxycarbonyl; alkyl-carbamoyl; or arylcarbonyl; or sulpho (HO_3S) ; phosphono ($(HO)_2P(O)$-) ; or a mono-, di-or tri-saccharide; wherein any alkyl group is optionally substituted with aryl, amino, mono-or di-alkyl-amino, a carboxylic acid residue (-COOH), or any combination thereof; and

r. Derivative forms of the above compounds as defined as items a to q, in which the 3-position carbon atom or, in the case of Formulae II and III, the 3-position carbon atom, the 26-position carbon atom or each of the carbon atoms at the 3-and 26-positions, carries an 0-sugar moiety wherein the sugar group is a mono-, di-or tri-saccharide, and acylated derivatives thereof.

3. (withdrawn) A use according to claim 1, wherein the active agent, or at least one of the active agents, is selected from compounds of the general formula Ia.

4. (withdrawn) A use according to claim 1, wherein the active agent, or at least one of the active agents, is selected from:

sarsasapogenin

sarsasapogenin cathylate

sarsasapogenin acetate

sarsasapogenin succinate and pharmaceutically acceptable salts thereof

sarsasapogenin glycinate and pharmaceutically acceptable salts thereof

sarsasapogenin alaninate and pharmaceutically acceptable salts thereof

sarsasapogenin valinate and pharmaceutically acceptable salts thereof

sarsasapogenin phenylalaninate and pharmaceutically acceptable salts thereof

sarsasapogenin isoleucinate and pharmaceutically acceptable salts thereof

sarsasapogenin methioninate and pharmaceutically acceptable salts thereof

episarsasapogenin

episarsasapogenin cathylate

episarsasapogenin acetate

episarsasapogenin succinate and pharmaceutically acceptable salts thereof

episarsasapogenin glycinate and pharmaceutically acceptable salts thereof

episarsasapogenin alaninate and pharmaceutically acceptable salts thereof

episarsasapogenin valinate and pharmaceutically acceptable salts thereof

episarsasapogenin phenylalaninate and pharmaceutically acceptable salts thereof

episarsasapogenin isoleucinate and pharmaceutically acceptable salts thereof

episarsasapogenin methioninate and pharmaceutically acceptable salts thereof

smilagenin

smilagenin cathylate

smilagenin acetate

smilagenin succinate and pharmaceutically acceptable salts thereof

smilagenin glycinate and pharmaceutically acceptable salts thereof

smilagenin alaninate and pharmaceutically acceptable salts thereof

smilagenin valinate and pharmaceutically salts thereof

smilagenin phenylalaninate and pharmaceutically acceptable salts thereof

smilagenin isoleucinate and pharmaceutically acceptable salts thereof

smilagenin methioninate and pharmaceutically acceptable salts thereof

epismilagenin

epismilagenin cathylate

epismilagenin acetate

epismilagenin succinate and pharmaceutically acceptable salts thereof

epismilagenin glycinate and pharmaceutically acceptable salts thereof

epismilagenin alaninate and pharmaceutically acceptable salts thereof

epismilagenin valinate and pharmaceutically acceptable salts thereof

epismilagenin phenylalaninate and pharmaceutically acceptable salts thereof

epismilagenin isoleucinate and pharmaceutically acceptable salts thereof

epismilagenin methioninate and pharmaceutically acceptable salts thereof.

saponin derivatives of sarsasapogenin, episarsasapogenin, smilagenin and epismilagenin in which, in each case, the 3-position carbon atom carries an 0-sugar moiety wherein the sugar group is selected from glucose, mannose, fructose, galactose, maltose, cellobiose, sucrose, rhamnose, xylose, arabinose, fucose, quinovose, apiose, lactose, galactose-glucose, glucose-arabinose, fucose-glucose, rhamnose-glucose, glucose-glucose-glucose, glucose- rhamnose, mannose-glucose, glucose- (rhamnose)-

glucose, glucose- (rhamnose)-rhamnose, glucose- (glucose)-glucose, galactose-(rhamnose)-galactose and acylated derivatives thereof; 16, 22-epoxycoprostan-3ß-ol, smilagenone, coprosterol, and pharmaceutically acceptable pro-drugs and salts thereof.

5. (currently amended) The method of claim ~~1~~9, wherein the active agent is present in a composition selected from pharmaceutical compositions, foodstuffs, food supplements and beverages.

6. (currently amended) The method of claim ~~1~~ 9, wherein the active agent is present with one or more additional active agents.

7. (currently amended) The method of claim 6, wherein the one or more additional active agent is selected from, but not limited, to cholinesterase inhibitors, dopamine agonists, COMT inhibitors, MAO-B inhibitors, anti-cholinergics, acetylcholine agonists, serotonin agonists, AMPA receptor agonists, GABA receptor agonists, NMDA receptor agonists, ß- adrenceptor agonists, digoxin, dobutamine, antiinflammatories, neurotrophic factors, statins, adenosine A2a receptor antagonists, aldose reductase inhibitors, immunomodulators, cannabinoid agonists, interferon (3 or tricyclic anti-depressants.

8. (cancel)

9. (new) A method for the treatment of amyotropic lateral sclerosis (ALS) comprising administering to a human or non-human animal an effective amount of an active agent, wherein the active agent is sarsasapogenin.